SUPERIOR CANAL DEHISCENCE SYNDROME – CASE REPORT

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ABSTRACT

We report a case of a patient with superior semicircular canal dehiscence syndrome, a recently described condition in which vestibular imbalance and/or hearing loss results from the discontinuity of the bone overlying the superior semicircular canals.

Case report. A 46-years-old women presented with autophony in the left ear and imbalance when shouting (Tullio phenomenon). Temporal bone computer tomography revealed a defect of the left superior semicircular canal caused by an enlarged superior petrosal sinus receiving drainage from a large cerebellar developmental venous anomaly.

Conclusion. We review superior semicircular canal dehiscence syndrome and its management, and we discuss common aetiologies. We conclude that superior semicircular canal dehiscence syndrome may present with a solely developmental aetiology, despite presenting late in life.

Key words: semicircular canal dehiscence, vertigo, hearing loss

INTRODUCTION

Superior canal dehiscence syndrome (SCD syndrome) is a rare medical condition of the inner ear, caused by a thinning or complete absence of the part of the temporal bone overlying the superior semicircular canal of the vestibular system. This may result from slow erosion of the bone or physical trauma to the skull and there is evidence that the defect or susceptibility is congenital. The SCD syndrome was first described by Dr Lloyd B. Minor in 1998. SCD syndrome can affect both hearing and balance, to different extents in different patients.

CASE REPORT

We present the case of a patient suffering of a sound-induced loss of balance, often associated with loud noises (Tullio phenomenon). The patient described a wide range of sounds that affect balance: a knock of the door, a telephone ringing and music. A change of pressure within the middle ear, for example when flying, caughing or nose-blowing, may equally set off a bunt of disequilibrium, known as the Hennebert sign. The patient also claimed a progressive hearing loss in the left ear.

The clinical examination started with the otomicroscopy which reveals a normal aspect of the tympanic membrane.

Acoumetry revealed a negative Rinne in the left ear and a lateralized Weber to the left ear.

Videonystagmography (VNG) revealed the absence of the spontaneous nystagmus; positive pressure within the middle ear (Valsalva) leads to a vertical-torsional nystagmus "going down", with the fast phase to the right ear, and vertigo.

Audiological evaluation included pure tone audiometry (PTA) which revealed mild low-frequency conductive hearing loss in left ear (Fig. 1) and

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impedancemetry. This showed bilateral type A tympanometry and bilateral normal acoustic stapedial reflex (Fig. 2) and Tullio phenomenon in acoustic stapedius reflex testing.

Due to history and audiological results we considered SCD syndrome as a possible positive diagnostic and we performed vestibular evoked myogenic potentials test (VEMP). Response (waves P1,

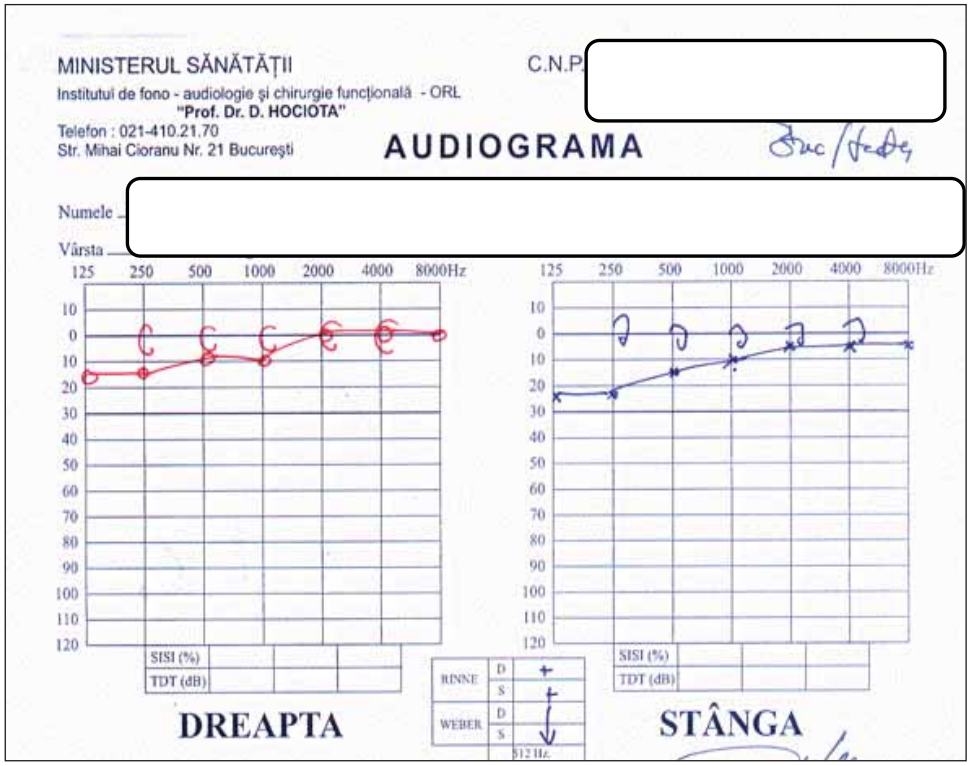

FIGURE 1. *PTA Left ear – mild low-frequency conductive hearing loss*

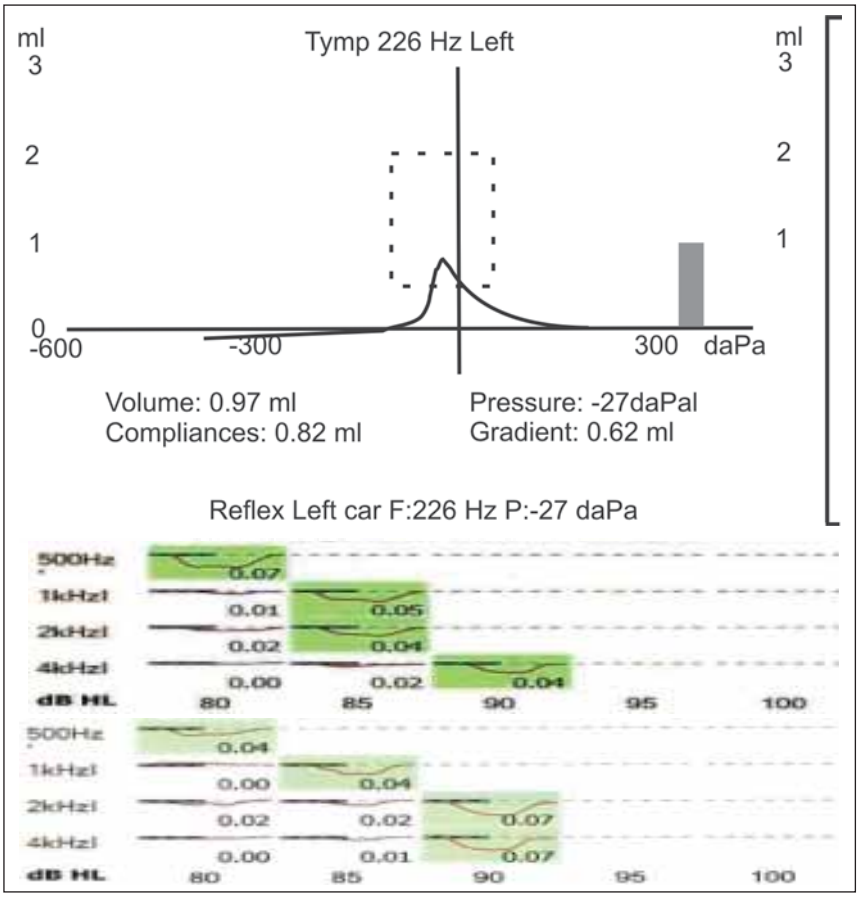

FIGURE 2. *Impedancemetry in left ear Type A tympanometry Normal acoustic stapedial reflex result (ipsi- and contralateral stimulation)*

N1) was present in both ears, but with significant amplitude difference between the ears, in favour of the left one. At the same acoustic intensity, VEMP response had huge amplitude in the left ear, three times larger than in the normal right ear (Fig. 3).

Based on these audiological measurements, we strongly suggest a SCD syndrome in the left ear and we referred the patient to computer tomography (CT) examination.

High definition coronal CT scan of the temporal bone is the most reliable way to distinguish between superior canal dehiscence syndrome and other conditions of the inner ear involving similar symptoms as Meniere disease and perilymphatic fistula.

In our case, the coronal CT scan revealed the erosion of the bone in the postero-superior part of the temporal bone, overlying the superior semicircular canal of the vestibular system (Fig. 4) due to an enlarged superior petrosal sinus receiving drainage from a large cerebellar developmental venous anomaly.

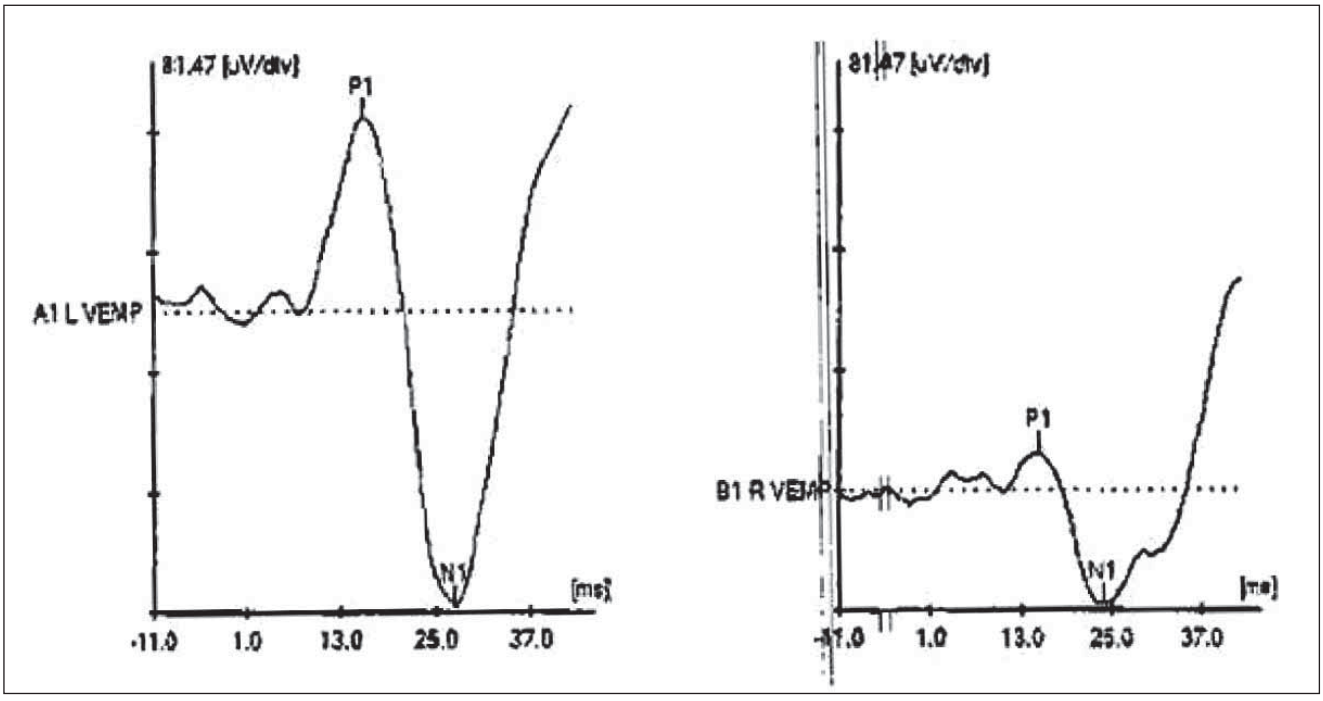

FIGURE 3. *VEMP recording – 97dB nHL click Left ear – high amplitude of the VEMP (3x right ear response)*

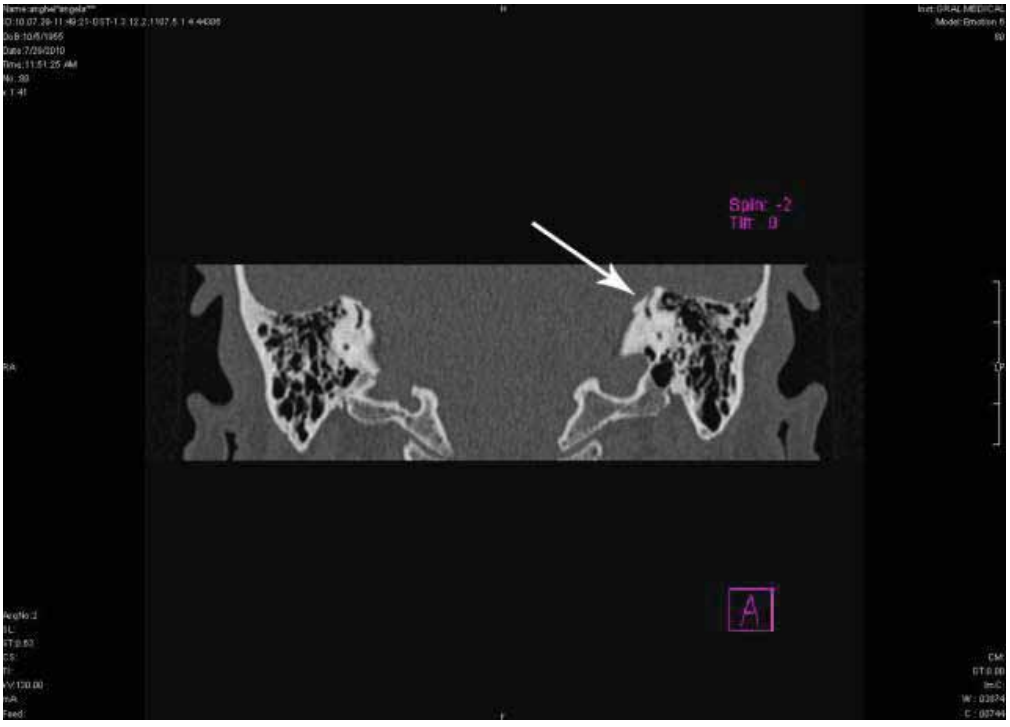

FIGURE 4. *Coronal CT scan (0.5-mm) of the left temporal bone dehiscence of bone (arrow) over the left superior semicircular canal*

An accurate history and non-invasive diagnosis is of great significance in SCD syndrome patients, as unnecessary exploratory middle ear surgery for conductive hearing loss may thus be avoided.

TREATMENT

From literature, when SCD syndrome is diagnosed, the osseous defect in the temporal bone can be repaired by surgical resurfacing of the affected bone or plugging of the superior semicircular canal. Bone cement has been the material most often used, in spite of its tendency to slippage and resorbtion, and a consequent high failure rate; recently, soft tissue grafts has been substituted.

Sometimes, prevention is enough for mild symptoms and our first choice of treatment was to recommend avoiding of the precipitating factors (stimuli) for three month period. If prevention would be insufficient, we shall refer the patient to neurosurgery evaluation.

DISCUSSION

Superior semicircular canal dehiscence syndrome is caused by abnormal communication between the inner ear and the cerebral middle fossa. Our paper review the clinical features and audiological and imagistic battery test recommended in patients with possible SCD syndrome.

Usually, SCD syndrome presents with pressure– or sound–related vertigo/imbalance (Tullio phenomenon and Hennebert sign, respectively) and mild hearing impairment or just autophony. The symptoms results from the osseous defect of bone covering the superior semicircular canal, exposing the vestibular system to variations of intracranial pressure and to bone-transmitted sound, resulting in so-called "third window" conduction. Most patients report noise- or pressure- related vertigo only on direct questioning, highlighting the importance of good history-taking in diagnosing superior semicircular canal dehiscence syndrome.

The otological examination revealed normal aspect of the tympanic membrane, with no membrane movements with respiration that should point clinicians away from Eustachian tube conditions and suggest superior semicircular canal dehiscence. There may be involuntary torsional eye movements and nystagmus relating to the affected canal when preesure varies in the external auditory canal. Investigating SCD syndrome requires PTA and impedancemetric evaluation of the hearing as well as high resolution (< 0.6 mm slice thickness) CT scanning to visualize the dehiscence.

The aetiology in our case is likely to be a developmental variation – an enlarged superior petrosal sinus receiving drainage from a large cerebellar developmental venous anomaly which slowly erodated the bone cover of the superior semicircular canal; dehiscence of bone overlying the superior semicircular canal generates a 'third window' for abnormal sound and pressure conduction.

The management of superior semicircular canal dehiscence syndrome involves conservative (preventing) and surgical approaches. Plugging of or resurfacing the dehiscence does lead to improvement or resolution of pressure or sound-related vertigo.

CONCLUSION

Superior semicircular canal dehiscence syndrome is a recently described disorder giving rise to vestibular and auditory symptoms. This pathology should be taken into account in differential diagnostic in patients with sound or pressure induced imbalance.

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